

## Unique Case of Atypical Central Neurocytoma with Craniospinal Metastases and Pituitary Adenoma

### Abstract

Atypical central neurocytoma with extracranial metastases is a rare variant of benign central neurocytoma (CN). No definitive course of treatment exists for atypical CN. We report a unique case of atypical CN with concomitant pituitary macroadenoma and subsequent metastases to the spine. The patient received craniospinal radiation therapy. Close-follow up post tumor-resection may be advised to monitor for drop metastases. To the best of our knowledge, this is the only case reported of atypical CN with drop metastases to the spine concomitant with pathological-proven pituitary macroadenoma.

**Keywords:** Drop, intraventricular, MIB, radiation, suprasellar

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### Introduction

Central neurocytomas (CN) are benign lesions that typically develop from the nuclei of the septum pellucidum and present as intraventricular tumors. CN is generally a benign tumor classified as Grade II by the World Health Organization and associated mainly with a favorable outcome.<sup>[1]</sup> Atypical CN can have aggressive nature of metastasizing to other areas of the brain and to extracranial areas, including the spine. Immunohistochemistry stain cell proliferation index (MIB-1) can help to differentiate atypical CN from benign CN. Here, we report a case of atypical CN that metastasized to the cervical, thoracic, and lumbar spine, with concomitant pituitary macroadenoma found on imaging. We believe this is the only case reported of this kind.

### Case Report

A 48-year-old male presented with 11-month history of syncopal episodes and questionable seizures. The patient was seen at an outside hospital and referred to our institution for neurosurgical care after computed tomography (CT) and magnetic resonance imaging (MRI) brain showed a mass centered in the trigone of the left lateral ventricle as well as pineal gland

extending into the third ventricle and fourth ventricle with local mass effect and possible infiltration of the left and right cerebral peduncles, left thalamus, and splenium of the corpus callosum [Figure 1a]. A suprasellar mass consistent with pituitary macroadenoma compressing optic chiasm was also noted [Figure 1b]. Enlargement of the lateral and third ventricles was consistent with obstructive hydrocephalus [Figure 1c]. Periventricular white matter signal changes with increased signal intensity on T2-weighted images were consistent with subependymal cerebrospinal fluid (CSF) migration [Figure 1c]. On presentation, he had normal vision and no papilledema with an unremarkable neurological examination. The working diagnosis preoperatively was pituitary tumor and hydrocephalus due to obstructive tumor in the left thalamopeduncular area.

The patient underwent an endoscopic biopsy and subtotal resection (STR) of the intraventricular tumor and ventriculostomy with a subsequent VP shunt. Intraoperatively, approximately 50% of the tumor, 6 cm, was removed from the left ventricle. Grossly, the tumor was hemorrhagic and had reddish consistency. Histologically, the tumor had a cellular neuroendocrine appearance with perivascular orientation and occasional pyknotic nuclei [Figure 1d]. Synaptophysin immunostain was diffusely positive,

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whereas glial fibrillary acidic protein (GFAP) stain was negative. MIB-1 proliferation rate was 5%–15%. These findings were consistent with the diagnosis of atypical CN. Pituitary mass was not operated on because of the lack of vision change, and therefore, was monitored until the patient developed vision changes.

The patient began radiation therapy targeting the pineal gland tumor. Due to concern for malignancy with the pineal gland tumor, MRI of the spine was taken 2 months after biopsy which showed no significant mass on the cervical spine but a focal enhancing lesion in the spinous process of T10 and curvilinear enhancement adjacent to the filum terminale at the S2 level. Repeat MRI of the thoracic and lumbar spine 3 months after the biopsy did not show significant changes. MRI brain 1-year postresection demonstrated no new evidence of recurrence of pineal tumor [Figure 2]. The patient was then lost to follow-up to oncology for about 1 year due to multiple missed appointments from hospitalizations. His headaches and syncopal episodes had resolved, however, he was having recurrent seizures and anomia.

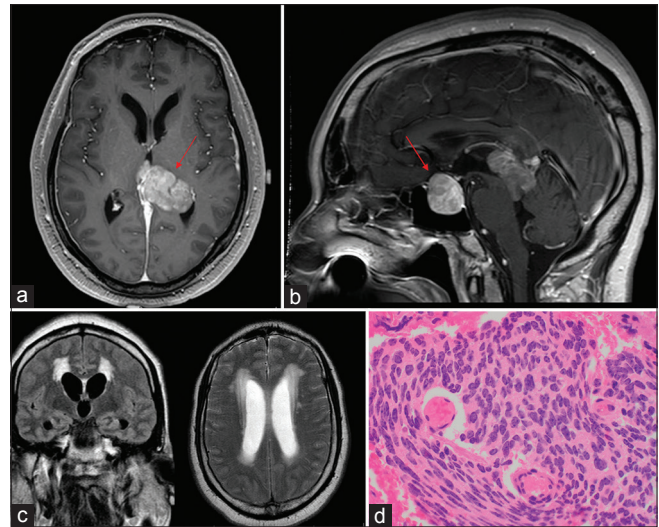
The patient presented to oncology approximately 2 years later with peripheral visual changes complaining of darkness over the right peripheral vision. On physical examination, the patient had decreased vision in the peripheral fields. CT brain showed an increase in size of suprasellar mass that was compressing the optic chiasm. Transsphenoidal pituitary tumor resection was performed; however, the fibrous nature of the tumor precluded complete resection. The surgical pathology report confirmed the morphologic and immunohistochemistry findings of the pituitary adenoma.

Repeat MRI brain 1 month after transsphenoidal pituitary tumor resection reported new enhancing lesions in the left cerebellum, medulla, and spinal cord, with an increase in the size of the pituitary macroadenoma [Figure 3]. The cranial approach was performed subsequently to resect more tumor. The postoperative CT brain showed unchanged right posterior parietal ventriculostomy shunt terminating at the septum pellucidum [Figure 4].

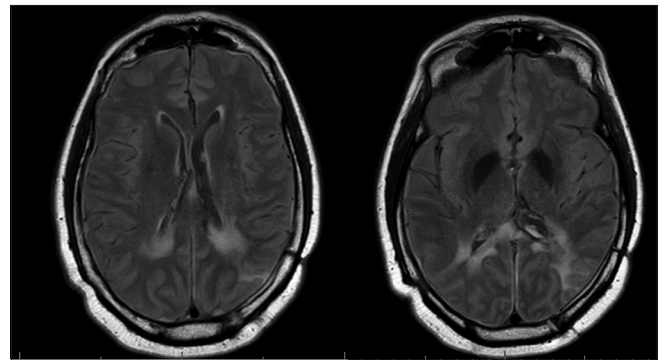
The radiologist recommended MRI spine to monitor for drop metastasis, because surveillance MRI brain showed a mass on the upper part of the cervical spine. MRI of the spine revealed extensive subarachnoid seeding throughout the cervical, thoracic, and lumbar spine [Figure 5]. The patient complained of leg weakness and had bilateral hemiparesis (3/5 strength). He was subsequently treated with craniospinal radiation therapy, temozolomide, and dexamethasone. He continued to have seizures despite anti-epileptic therapy and progressive worsening of gait instability. He was transitioned to hospice care.

## Discussion

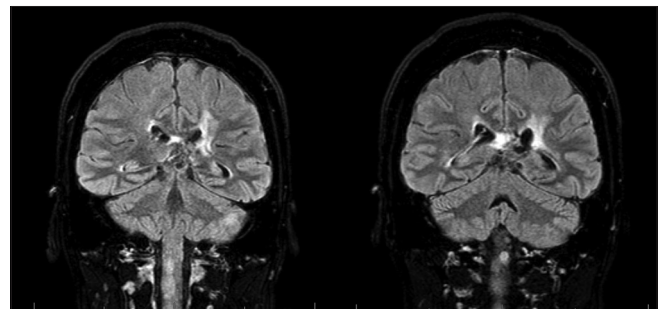
CN was first described by Hassoun *et al.*, in 1982 as rare intraventricular brain tumors, comprising only



**Figure 1:** (a) Magnetic resonance imaging brain axial T1 postcontrast. Central neurocytoma on the left lateral ventricle. (b) Magnetic resonance imaging brain sagittal T1 contrast three-dimensional. Pituitary macroadenoma. (c) Magnetic resonance imaging brain coronal T2 fluid-attenuated inversion recovery (left) and axial T2 (right). (d) High-power H and E of tumor. Neuroendocrine appearance with perivascular orientation



**Figure 2:** Magnetic resonance imaging brain axial T2 fluid-attenuated inversion recovery



**Figure 3:** Magnetic resonance imaging brain coronal T2 fluid-attenuated inversion recovery fat suppression

0.25%–0.5% of all brain tumors.<sup>[2,3]</sup> Both sexes are equally affected by this tumor, with a median age at the time of diagnosis was 34 years.<sup>[4]</sup> The typical clinical presentation of CN includes headache, memory or vision problems, seizures, vomiting, nausea, and increased intracranial pressure, which can lead to hydrocephalus.



Figure 4: Computed tomography brain axial

Most neurocytomas develop from the nuclei of the septum pellucidum.<sup>[5]</sup> Radiological imaging and pathological finding from a biopsy confirm the diagnosis. Immunohistochemistry is performed for the diagnosis of CN because of its histological similarities to brain tumors such as ependymomas and oligodendrogliomas. Positive staining for synaptophysin is a major indicator of the presence of CN. The absence of other markers, including GFAP, neurofilament, Vimentin, and neuron-specific enolase, aid in confirming the diagnosis of CN.<sup>[6,7]</sup>

Gross total resection (GTR) of the tumor is stated as the best course of treatment and favors survival rates without recurrence.<sup>[8]</sup> While CN is generally associated with a favorable prognosis, more aggressive clinical cases are known. Such cases are termed atypical CN and can be characterized by recurrence and atypical histological features such as increased mitotic activity or vascular proliferation.<sup>[9,10]</sup> Atypical CN with malignant behavior has shown an increased tendency to disseminate through the CSF causing spinal metastasis.<sup>[11-14]</sup> MIB-1 labeling index is a key marker in determining the prognosis of CN. An MIB-1 labeling index of  $\geq 2\%$  or  $\geq 3\%$  is a defining feature of atypical CN and portends a poorer survival rate and a higher risk of relapse.<sup>[15,16]</sup> In general, when GTR is not an option, STR of the tumor is performed. STR has a 3.8 times higher risk of adverse outcomes than GTR.<sup>[8]</sup> Adjuvant radiotherapy or chemotherapy has been stated as additional courses of treatment for patients presenting with atypical CN. Since no definitive course of treatment exists for atypical CN, case reports provide anecdotal experience in treating this rare metastatic lesion.

## Conclusion

Our patient was a unique case of atypical CN with drop metastases to the spine because he had a pathological-proven pituitary macroadenoma concomitant with the unrelated atypical CN. The patient had lost follow-up to oncology for almost 1½ years during which the patient developed

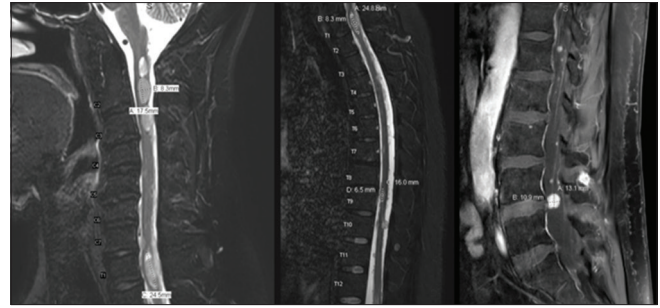


Figure 5: Magnetic resonance imaging C-spine sagittal short tau inversion recovery (left). Magnetic resonance imaging T-spine sagittal short tau inversion recovery (middle). Magnetic resonance imaging L-spine sagittal T1 fat suppression (right)

drop metastases to the spine causing bilateral lower leg weakness and neck pain. Close follow-up, therefore, is prudent for patients with atypical CN to detect earlier further metastases. In our patient, it was the worsening vision due to the pituitary macroadenoma that led to prompt brain imaging, which then revealed spinal metastases to the upper cervical cord.

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## Conflicts of interest

There are no conflicts of interest.

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